International Referral and Elderly Care—A Case of Atypical Parkinsonism and Cerebellar Atrophy

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SUMMARY

A 66-year-old male patient had a history of hepatitis B, was a hepatitis C carrier, and had hypertension. He was referred to our family medicine international clinic by the Canadian Physician's Referral Service. He had had progressive general weakness over 2 years, and the symptom had recently exacerbated. He also had slurred speech, difficulty in swallowing with occasional choking, constipation, and urinary incontinence. He was diagnosed as having Parkinson disease, but his symptoms worsened despite treatment with levodopa 250 mg four times daily for 2 years. He had sought medical help in many clinics in Taiwan and Canada and was referred to our outpatient clinic by his Canadian family physician. The neurologist suggested that the diagnosis was multiple system atrophy (MSA) after history taking and neurologic examination. Rehabilitation programs, including physical therapy, occupational therapy and speech therapy, were arranged for him. Many Taiwanese immigrants prefer to come back to Taiwan for medical treatment. If we can integrate medical resources not only between different hospitals but also between different countries, many unnecessary medical expenses could be avoided. In this case, we summarized the case history and provided him with a CD of the images. This will be helpful in further care by a Canadian physician. [International Journal of Gerontology 2009; 3(3): 192–194]

Key Words: multiple system atrophy, parkinsonism, referral

Introduction

Parkinsonism is defined as the progressive reduction in speed and amplitude with repetition of limb movements. However, some parkinsonism is also associated with autonomic dysfunction and shows poor levodopa response. For such atypical parkinsonism, multiple system atrophy (MSA) may need to be considered. MSA is defined as a sporadic, adult-onset, progressive, neurodegenerative disease of undetermined etiology. The MSA patient manifests parkinsonism, pyramidal, cerebellar, and autonomic dysfunction in any combination. MSA is rarely recognized in clinical practice and is frequently mistaken for Parkinson disease even by a neurologist.

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Case Report

A 66-year-old male patient had a history of hepatitis B, was a hepatitis C carrier, and had hypertension. He was referred to our family medicine international clinic by the Canadian Physician's Referral Service. He had had progressive general weakness over 2 years, and the symptom had recently exacerbated. He also had slurred speech, difficulty in swallowing with occasional choking, constipation, and urinary incontinence. He was diagnosed as having Parkinson disease, but his symptoms worsened despite treatment with levodopa 250 mg four times daily for 2 years. He had sought medical help in many clinics in Taiwan and Canada and was referred to our outpatient clinic by his Canadian family physician.

At the outpatient clinic, the results of the physical and neurologic examinations showed clear consciousness with normal judgment, orientation, memory, abstract thinking, and calculation. The patient had a mask-like face with dysarthria and dysphagia. He had

a decreased deep tendon reflex in the lower limbs with cogwheel rigidity. He did not have a resting tremor but had an intention tremor discovered by the finger-nose-finger test. The finding of the Romberg test was positive and he had poor balance on standing. Ambulation was possible with the help of a walking frame, but he tilted to the left side. He did not have any significant laboratory abnormality. The brain magnetic resonance images showed disappearance of the pars compacta of the substantia nigra, and technetium Tc 99m TRODAT cerebral dopamine transporter single photon emission computed tomography images revealed impaired uptake in the bilateral basal ganglionic regions (Figure). The imaging findings were consistent with Parkinson disease. Under the impression of atypical Parkinson disease and cerebellar atrophy, we requested a neurologic consultation.

After examining the patient, the neurologist suggested that the patient had MSA. Rehabilitation was arranged for posture protection and therapeutic exercise. He also received speech therapy for verbal training and learned safe-swallowing skills. He was scheduled for a home visit after being discharged from our hospital.

Discussion

MSA is defined as a sporadic, adult-onset, progressive, neurodegenerative disease of undetermined etiology. The MSA patient manifests parkinsonism and pyramidal, cerebellar and autonomic dysfunction in any combination. Its prevalence is reported to be between 1.9 and 4.9 cases per 100,000 population[1–4]. MSA is rarely recognized in clinical practice and is frequently mistaken for Parkinson disease even by a neurologist[5]. The pathologic findings are cell loss, gliosis, and glial cytoplasmic inclusions in several brain and spinal cord structures. When autonomic failure predominates, MSA is sometimes termed *Shy-Drager syndrome*[6], but when extrapyramidal features predominate, the term *striatonigral degeneration type*, *parkinsonian variant*, or *MSA parkinsonism* is sometimes used. If cerebellar features predominate, MSA is sometimes termed *sporadic olivopontocerebellar atrophy* or *MSA cerebellar type*.

The clinical diagnosis of MSA that is now widely used was developed by the International Consensus Conference promoted by the American Academy of Neurology[7]. The consensus criteria specified three diagnostic categories of increasing certainty: possible, probable, and definite (Table). The diagnosis of probable MSA requires the criterion of autonomic and/or urinary dysfunction and the presence of poorly-levodopa-responsive parkinsonism or cerebellar ataxia. Only pathologic findings can confirm the diagnosis of MSA. Generally, MSA patients do not have a robust response to medications for Parkinson disease and its clinical course progresses faster than Parkinson disease.

This patient had emigrated from Taiwan to Canada. His family physician in Canada referred him back to Taiwan for further evaluation and treatment during a visit to friends in Taiwan in March 2008. His family sent him to many clinics in Taiwan because of the poor response to medications for Parkinson disease. The family doctor's role is to integrate the medical information

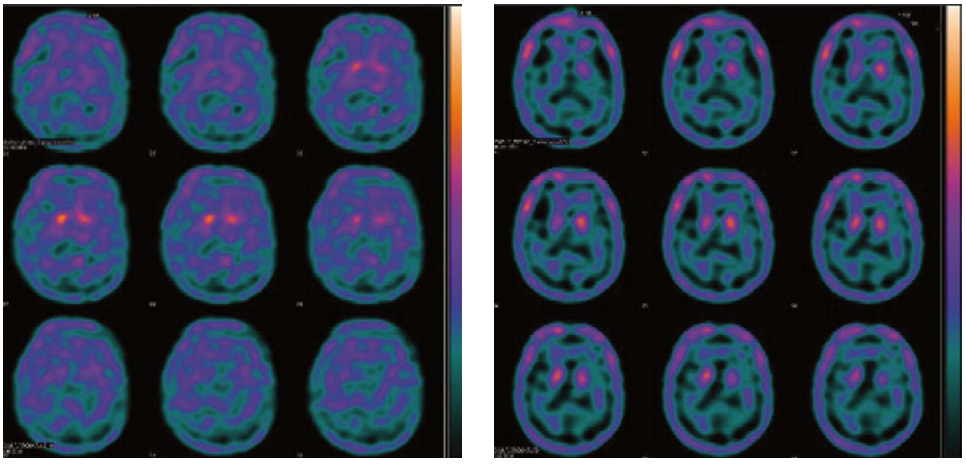

Figure. Technetium Tc 99m TRODAT cerebral dopamine transporter single photon emission computed tomography images revealed impairment of uptake in the bilateral basal ganglia regions. The radioactivity uptake in bilateral basal ganglia showed normal caudate but no putamen uptake. The image findings were compatible with parkinsonism.

Table. *Multiple system atrophy (MSA): clinical diagnosis guidelines of the International Consensus Conference**

Feature	Defining criteria	Disease characteristics
Autonomic failure	(1) Orthostatic fall in blood pressure by > 30 mmHg (systolic) or 15 mmHg (diastolic) (2) Persistent urinary incontinence with erectile dysfunction (in men) (3) Both (1) and (2)	Orthostatic hypotension Urinary incontinence Bladder-emptying difficulty Erectile dysfunction
Parkinsonism	Bradykinesia plus (1) rigidity, (2) postural instability, or (3) tremor	Progressive reduction in speed and amplitude with repetition of limb movements Rigidity Primary postural reflex loss Tremor (postural, rest, or both) Minimal or transient response to L-dopa or other dopaminergic drugs
Cerebellar dysfunction	Gait ataxia plus (1) limb ataxia, (2) sustained gaze-evoked nystagmus, or (3) ataxic dysarthria	Wide-based stance and irregular steps Limb ataxia Ataxic speech Gaze-evolved nystagmus

**Possible MSA: one criterion plus two disease characteristics from other domains; probable MSA parkinsonism: autonomic criterion plus poorly-responsive parkinsonism; probable MSA cerebellar type: autonomic criterion plus cerebellar dysfunction; definite MSA: pathologic confirmation.*

and provide professional opinion and continuing care to the patient[8]. In this case, we summarized the case history and provided the images on CD to him. This will be helpful for further care by a Canadian physician.

Medical costs are lower in Taiwan than in Western countries[9]; therefore, many emigrants tend to return to Taiwan if medical needs arise overseas. If the medical reports and images provide detailed information between different hospitals in various countries, many unnecessary medical expenses can be avoided.

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